261

Pulmonary estimation after liver transplantation in CF children

H. Dmenska[1], P. Gutkowski[1], P. Kalicinski[2], M. Markiewicz[2]
[1]Lung Physiology Dept,[2]Dept of Pediatric Surgery and Organ Transplantation, The Children's Memorial Health Institute, Warsaw

Long-term survival in CF has led to revealing extrapulmonary complications including biliary cirrhosis (fatal in up to 5% pts prior to the advent of liver transplantation (LTx)). It causes increased morbidity and mortality and LTx is the only effective therapy. Aims: Description of the clinical and pulmonary characteristics of CF pts considered for LTx. Methods: From 1990 to 2004, 215 pts received 241 LTx, of these 4 with CF. Records of all CF pts with LTx were reviewed. Results: 4 pts with CF were the recipients of 5 liver allografts. Mutation analysis showed: ΔF508/ΔF508, ΔF508/G542X and ΔF508/R553X. Two LTx were cadaveric, and two from related donors. Mean age at LTx was 9.9 yr (range 7.6-13.7). Mean survival time was 1.7 yr (range 0.2-3.6). Pt 3 who underwent retransplant for primary allograft nonfunction died of systemic multiorgan failure. Immunosuppression was based on routine medicines. Pulmonary function tests (PFT) were obtained one year before LTx in 4 and post LTx in 3 pts.

	Pre LTx [% pred]			Post LTx [% pred]		
Patients No	VC	FEV_1	FEF_{50}	VC	FEV_1	FEF_{50}
1.	82	91	104	78	85	105
2.	95	104	103	112	125	96
3.	107	103	59	-	-	-
4.	65	64	50	69	69	57

There was no significant deterioration in posttransplant PFTs. All pts had had positive sputum cultures before LTx including Staphylococcus aureus (Sa) (4 pts) and Pseudomonas aeruginosa (Pa) (2 pts) and postoperatively Pa (3 pts) and Sa (1 pt). Conclusions: LTx is the acceptable treatment for CF pts with mild to moderate pulmonary disease and end-stage liver disease. In our pts PFT improved slightly or remained unchanged. Immunosuppressive agents did not have a deleterious effect on pulmonary function. LTx in carefully selected CF pts offers opportunity to reduce morbidity and to increase quality of life.

262

Relationship between impulse oscillometry (IOS) and spirometric indices (SI) in cystic fibrosis (CF) children

L. Moreau[1], D. Crenesse[1], F. Berthier[2], M. Albertini[1]
[1]Laboratoire d'Explorations Fonctionnelles Respiratoires Pédiatriques, Hôpital de l'Archet II, Nice, France, [2]Département d'information et d'informatique médicale - Hôpital de Cimiez, Nice, France

The aim of our retrospective study was to determine the relationship between IOS data and spirometric tests in CF children. Thirty CF children aged 4-19 years have performed lung function tests (LFT) in a fixed sequence (IOS → Spirometry) on 1 to 7 separate occasions. IOS parameters were respiratory resistance, reactance and impedence at 5 Hz (R5, X5, Zr) and the resonant frequency (Fres). SI included FEV1, $MMEF_{25-75}$ and FVC. An inverse relationship was observed between raw values of R5, Zr, Fres and SI respectively, and X5 correlated positively with these SI. Although significant, these correlations were poor. Receiving Operating Characteristic curves were constructed to identify cut-off points for IOS parameters to discriminate between children according to different FEV1 thresholds (percent of predicted values), generally used to describe the severity of lung function impairment as following : normal, FEV1 ≥ 80% ; mild, FEV1 ≥ 60% ; moderate, FEV1 ≥ 40% ; severe, FEV1 < 40%. The results are exploitable only for the two first thresholds given the small number of patients for the lowest threshold. No acceptable cut-off points can be found for IOS parameters. Analysis of trends were carried out on a subset of 15 patients who repeated LFT on at least 5 separate occasions. Contrary to the time course of FEV1 who showed a significant diminution, none of the IOS indices demonstrated a consistent trend, and there was no correlation between the evolution of FEV1 and that of each IOS parameter. So, IOS measurements presented an insufficient sensitivity to detect and follow bronchial obstruction in CF patients.

263

The assessment of respiratory status in patients with cystic fibrosis: bodypletysmography or impulse oscillometry?

A. Minarowska[1], M. Kaczmarski[1], Ł. Minarowski[3], M. Mrugacz[2]
[1]IIIrd Department of Children's' Disease, Medical University of Bialystok Poland; [2]Department of Pediatric Ophthalmology, Medical University of Bialystok, Poland; [3]Medical University of Bialystok, Poland

Background: The progress of studies over the ventilation mechanisms along with dynamic development of novel measurement techniques enabled using this method for examination also in patients with cystic fibrosis (CF). Application of appropriate technique of measurement is important to asses the advancement of the destruction of lung tissue.
Aim: The aim of the work was to assess the resistance in airway using bodypletysmography and impulse oscillometry (IOS) in group of patients with CF.
Material and methods: The study was performed on group of 18 patients with CF (7F, 11M) aged 10-27 yrs (mean age 15,4 yrs) and on group of 45 healthy patients (25F, 20M) aged 9-24 yrs (mean age 13,2 yrs). In both groups bodypletysmography and IOS were performed using Jaeger MasterLab and IOS MasterScreen apparatus. Total airway resistance (Rtot) and resistance at frequency of 5Hz (Rat5Hz) were assessed in both groups. Statistical analysis was conducted subsequently.
Results: Obtained mean values were respectively: Rtot = ,4 ± 0,26 kPa/l/s, Rat5Hz = 0,58 ± 0,32 kPa/l/s (max. = 1,31 kPa/l/s) in studied group and Rtot = 0,25 ± 0,08 kPa/l/s, Rat5Hz = 0,5 ± 0,21 kPa/l/s (max. = 0,91 kPa/l/s) in controls. Correlation coefficient of Rat5Hz and Rtot was r=0,8 (p<0,001) in patients with CF.
Conclusions: Novel examination techniques of airway resistance assessment, like impulse oscillometry, and bodypletysmography are complementary, but in burdensome situations IOS has more advantages than bodypletysmography in assessing the airway status in patients with CF.

264

Sleep hypoxemia in stable cystic fibrosis(CF)

M. Kakoura[1], E. Fouka[1], M. Fotoulaki[2], D. Chloros[1], G. Bompotis[3], E. Kotsi[2], S. Vogiatzis[1], S. Zanos[1], S. Nousia-Arvanitakis[2]
[1]Lung Unit, [2]4th Dept of Pediatrics Aristotle's University of Thessaloniki, [3]Dept of Cardiology, General Hospital "PAPAGEORGIOU", Thessaloniki, Greece

Background: Sleep hypoxemia due to hypoventilation, occurring predominantly in REM sleep, is the main etiologic factor in the development of cor pulmonale in patients with CF. It is important to predict sleep-related desaturation/ hypoxia as this could influence the timing of initiation of preventive treatment and minimizing the load on cardiovascular system.
Purpose: To identify predictors of sleep hypoxemia in CF patients.
Methods: We correlate sleep-occurring hypoxia and hypopneas/apneas index to daytime measurements of lung function, respiratory muscle strength, arterial blood gases, echocardiographic measurements and subjective estimation of sleep quality in 25 clinically stable adolescent and adult CF patients (13 M, age 20±6) having mild to moderate lung disease. They were administered with the Pittsburgh Insomnia Scale. Unattended sleep studies with the Embletta portable system were performed on the same night. All patients underwent PFTs (lung volumes, flow rates and diffusing capacity for carbon monoxide), maximal Inspiratory (P_{Imax}) and Expiratory muscle (P_{Emax}) pressure measurements, echocardiographic and arterial blood gases evaluation.
Results: Overnight oximetry demonstrates that mean SpO2 and DLCO% pred were highly correlated (r=-0.619, p < 0.001), as well as min SpO2 and left ventricle end diastolic diameter(r=-0.465, p<0.02). No correlation was found between subjective sleep quality and physiologic variables describing disease severity.
Conclusions: A decrease in lung diffusing capacity for carbon monoxide correlates with sleep hypoxemia. In such cases an overnight oximetry might help to reveal sleep hypoxemia at an earlier stage.